

MELANOTIC SARCOMA OF THE COMMON BILE
DUCT AND THE AMPULLA OF VATER.*

BY FRANCIS J. SHEPHERD, M.D., F.R.C.S., EDIN. (HON.)

OF MONTREAL,

Surgeon to the Montreal General Hospital.

THE rarity and obscurity of this case constitutes my apology for reporting it.

Mr. A. S., aged 44, bridge engineer, consulted Dr. A. E. Morphy, of Lachine, P. Q., in June, 1907, for severe itching all over the body, but especially about the chest and back. On examination a very faint tinge of yellow was observed in the skin and conjunctivæ. There was no digestive disturbance but distinct loss of appetite. A few days later the jaundice deepened and the itchiness increased. There was no pain, but the patient complained of loss of strength, great lassitude and much depression of spirits. There was also constipation. On physical examination, when first seen by Dr. Morphy, no abnormal condition about the liver was discovered; everything seemed normal.

About the middle of July a smooth, rounded, painless swelling was made out in the region of the gall-bladder. This was looked upon as a distended gall-bladder. All this time there was abundant bile in the urine and the stools were colorless and very fetid, except occasionally when they had a chocolate hue.

Mr. S. was admitted under my care as a private patient in the Montreal General Hospital, July 26, 1907, the following being his history: A tall, spare man, of very fair complexion; had never been ill before in his life and had always been most athletic, being very fond of outdoor sports. He was in his usual health until May 15, 1907, when his feet began to itch at night. This itching spread all over his body and very soon (early in June) his conjunctivæ became yellowish in color and then his whole skin became jaundiced. The color deepened rapidly, and at the time this note was made his color was more yellow than it had previously been, the itching was less troublesome but he complained

* Read before the American Surgical Association, May 5, 1908.

of great depression and weakness. He never had any severe pain in the abdomen, nor headache, but complained of a sensation as if a rope was being tied about his abdomen below the costal margin accompanied by a dull ache, especially severe when he took a deep breath.

Examination showed that his lungs and heart were normal. The urine contained bile but no sugar or albumin; the bowels were constipated, stools foul and colorless. Has never had any vomiting or fever. Has lost about 35 pounds since illness commenced. Pulse slow, 58. On examination of the liver it was found to be slightly enlarged and a fulness was felt in the region of the gall-bladder, but no distinct tumor was made out.

A question arose as to the cause of this condition and it was felt that it was either due to gall-stone in the common duct or malignant disease. An exploratory operation was advised, and on July 30th the abdomen was opened by a vertical incision through the middle of the right rectus abdominal muscle. The gall-bladder was found fairly distended but no stone could be felt, nor could any nodules or enlarged glands be made out in the common duct, nor was any thickening or infiltration felt. The head of the pancreas was somewhat hard, but nothing definite was discovered, so the gall-bladder was opened and a lot of thick, dark colored bile evacuated mixed with mucus and a dark substance of the consistence of mud. A drainage-tube was left in and nothing more done. My conclusion was, from having had a somewhat similar case the previous year, that a soft secondary malignant growth was blocking the common duct below the junction of the cystic duct, but on careful examination no primary growth could be discovered anywhere. The rectum was normal and apparently all other parts also. It was a puzzling case and explanation was difficult. The tube discharged much bile and his jaundice cleared up considerably, but he continued getting weaker and was more difficult to feed.

A month after the operation his left parotid began to enlarge and soon after pus came from the left ear; a large abscess formed which was opened and discharged freely and much necrosed tissue was taken away. Another abscess formed in the submaxillary region and caused a considerable increase of his debility. He finally died on September 6, 1907, of exhaustion.

An autopsy was allowed and the following is an abstract of the findings as reported by the Pathologist of the Hospital, Dr. Duval.

Autopsy performed fourteen hours after death. In the region of the gall-bladder there is a linear scar 10 cm. in length parallel to the median line. Through the scar runs a fistulous tract which communicates with the gall-bladder and on pressure discharges bile. The inner surface of the gall-bladder appears normal. The cystic duct, opened in situ, is distorted into a shape like the letter S, apparently the result of old adhesions. The wall of the cystic duct is of normal thickness and its inner surface smooth throughout. On opening the common duct it is seen uniformly dilated to three times its normal calibre. Its wall is somewhat thinned and the rugæ of the tunica propria present a marked fenestration, due probably to long continued distention. In the lower portion of the duct there is a soft, brownish-black, fungoid mass, 2.5 cm. in length, which completely occludes the lumen of the common duct and part of Vater's diverticulum. The passage-way to the duodenum, however, is patent and normal. The tumor throughout its extent is intimately attached to the duct wall and appears to have started in the tunica propria; there is no thickening of the wall. The attachment of the growth maintains an even line 2 mm. within and along the wall as indicated to the naked eye by the pigment limit. (Fig. 1.)

The tumor is confined entirely to the common duct and ampulla. Neither the pancreas nor its duct are involved in the growth. There is no similar mass found elsewhere nor were any cutaneous moles found anywhere in the body. The common duct was opened in situ over a grooved director to which on passing it down the lumen no resistance was offered, so no occlusion was detected by means of the probe. The growth was so soft that only after the duct had been laid open was the obstruction discovered. The brownish-black mass now assumed a distinct cylindrical form which bulged far over the edges of the opened duct. It was only with great difficulty it could be returned to its original place and the edges of the duct brought again into apposition.

At first it seemed as if the tumor was altered blood or inspissated bile; its true nature was discovered, however, after a more careful examination. The melanoma on closer inspection showed

FIG. 1.

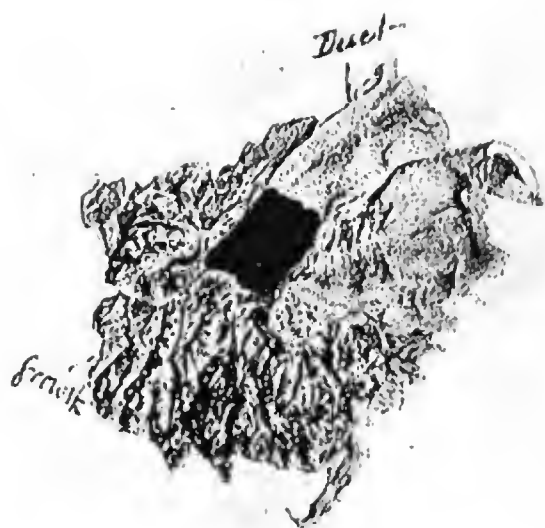
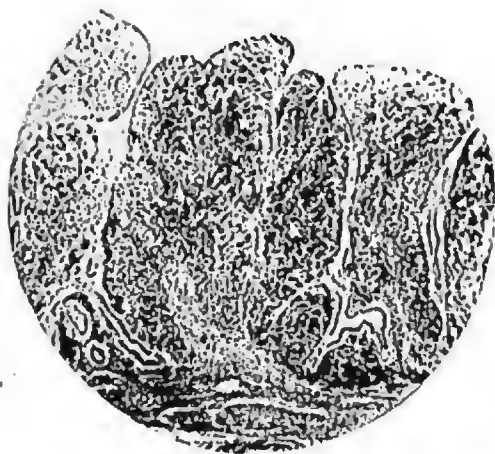


FIG 2



Section showing pigment cells.

FIG. 3.



Section showing villi.

innumerable densely arranged, flattened, finger-like projections; these floated free from one another at their distal extremities, but remained firmly attached at their base. When the mass was submerged in water the upper and lower limitations of the growth are sharply defined. The tumor with its curious villous structures resembles certain forms of vegetable algæ growing under water. It is noteworthy that no part of the mass could be washed away or the water discolored by the coloring matter of the tumor. On removing the mass from the water the villi immediately collapsed, allowing the tumor to assume again a smooth, dull, black, velvety surface.

Microscopical examination showed the growth to be composed almost entirely of pigmented cells. The pigment is most marked on the outer extremities of the villi where the alveolar structure is most pronounced. The cells have a large, distinctly lobulated vesicular nucleus with one or more nucleoli. The nucleolus is always sharply defined and may be very large, often only a narrow rim of nucleus surrounding an enormous nucleolus. When the cell contains little pigment it is small and the nucleus proportionately diminished. The pigment invariably occurs in the form of globules arranged in cell protoplasm equi-distant from one another. (Figs. 2 and 3.)

The pigment differs from that of the melano-sarcomata in that instead of the irregularly scattered masses of the latter it occurs in the form of globules evenly arranged within the cells. The distribution is so regular that it does not seem to be a by-product but rather an integral part of the cell. An occasional small vessel in the submucosa of the common bile duct in close proximity to the tumor contains pigment cells and with this exception the melanotic cells are strictly confined to the main mass. There was no metastasis found elsewhere in the body.

The region below the involved part of the duct showed a well-marked periglandular infiltration of lymphoid and plasma cells, but no pigmented cells of any description. All the gland follicles in the tunica propria above and below the growth showed a low grade of chronic peri-inflammation though other tissues were normal. The villous masses comprising the tumor are covered by a thin, though well-defined connective tissue, enveloped in whose fibrils are elongated cells arranged end to end in unbroken chain. There was no pigment either intra- or extra-cellular in this supporting tissue and in no way does it resemble melanoma.

Remarks.—The situation of this growth is unusual for a pigmented one, which is apparently primary and whose cellular structure resembles in many ways epithelioma. Secondary melanosarcomata are common in the liver, but no case has

been reported of such a secondary growth in the common bile duct. The gall-bladder and ducts are the more frequent sites for primary sarcoma. We must conclude from the absence of primary growth elsewhere that the melanotic growth arose from cells in the common bile duct.

There is a possibility that at some time there was a dislodgement of one or more pigment-bearing cells from a normal situation which subsequently became arrested at a distant point. In this way we may account for the occurrence of secondary melanoma in an internal organ where no primary tumor actually exists. Of course this theory is open to the criticism that the primary focus had escaped discovery.